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Fibrous Dysplasia

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If pathologic manifestations, which at first seem to be totally disconnected, are found together in a sufficient series of patients, some relation between them is apparent.[2]

Fibrous dysplasia is a disorder of expanding benign fibro-osseous lesions occurring in one, several, or multiple bones of the skeleton. The spectacular association of skeletal lesions with cutaneous pigmentation and endocrine dysfunction generated the earliest excitement and curiosity regarding this disorder.[2, 29] Clinical, pathological, and radiological comparisons and confusion were frequent with enchondromatosis, neurofibromatosis, and hyperparathyroidism. Lichtenstein[26] defined fibrous dysplasia as a distinct entity and with Jaffe[27] delineated the clinical manifestations into monostotic, polyostotic, and extraskeletal forms. Although subsequent reviews have enlarged our perspectives regarding the natural history of fibrous dysplasia, the etiology remains obscure.[17, 22–24, 34, 40] This disorder appears to be a developmental abnormality of bone forming mesenchyme.[27] The structural integrity of the affected bone segment is violated with fracture and deformity a common end result. The lesions of fibrous dysplasia begin in childhood, but recognition may not occur until adult life. No consistent familial or hereditary factors have been established. Recent interest in fibrous dysplasia has centered upon the pathogenesis of the endocrinopathies of the Albright syndrome and the increasing frequency of malignant transformation occurring in an area of fibrous dysplasia.[5, 10–12, 14–16, 20, 21, 24, 28, 30]

CLINICAL SYNDROMES

The diverse clinical presentations of fibrous dysplasia have been conveniently grouped into three categories: monostotic, single bone in-

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volvement; polyostotic, multiple bone involvement; and Albright's syndrome and its variants. Within each of these categories there can be enormous variations. The patient with a single localized lesion of the humerus or femur differs greatly from the patient wherein the entire humerus or femur is involved. However, both patients are classified as having monostotic involvement. It is not uncommon with monostotic fibrous dysplasia to discover distant skull or rib lesions many years after the original diagnosis. Polyostotic involvement can encompass two or more bones in the same extremity (monomelic distribution), a single extremity lesion and corresponding incidental lesions in the hemipelvis, or diffuse skeletal involvement with or without endocrine changes. The classification of fibrous dysplasia into these convenient groupings is helpful, not in numerical, but in a behavioral sense. Each category has a separate pattern of expression that allows us to anticipate management programs. Fibrous dysplasia manifests a continuum of skeletal involvement, and any classification is arbitrary at best.

Monostotic Lesions

The monostotic lesion would seem to be the most common form of fibrous dysplasia, especially if we consider that many monostotic lesions remain quiescent throughout an individual's life span. These asymptomatic lesions may be recognized only on an incidental radiograph or when a complication occurs, i.e., fracture or malignant transformation. Although any bone of the skeleton may be affected, the most common sites of monostotic involvement are the proximal femur, tibia, rib cage, and facial bones. Fibrous dysplasia is the commonest cause of a benign expansile lesion of the rib cage. The age range affected is from 10 to 70 years with the most frequent period of recognition in the second and third decades.[40] The age distribution is slightly older in this form than in polyostotic fibrous dysplasia because of the less severe clinical manifestations. There is no sexual predominance, although some authors report a female to male ratio as high as 3 to 1.[25] The more polyostotic cases contained in a review, the more female bias will be present in the sex distribution curves.

The clinical features of a solitary lesion are pain, fracture or deformity, or an enlarging mass. It is our impression that the dysplastic process in itself is not painful, but that the complications of the lesions cause the pain. The presenting symptom relates to the location of the lesion. Fracture through an attenuated cortex occurs in the extremities, whereas the pressure of an enlarging mass in the skull gives rise to headaches and increasing facial deformity. Over half of the rib lesions are asymptomatic. The cranial lesions may remain silent, but frequently are observed to progress slowly in adults toward visual impairment or hearing loss as a result of compromise of the optic nerve in the optic canal or stenosis of the external canal or middle ear by temporal bone changes, respectively.[36] Lesions often involve the long bones, cause no pain, apparently stabilize with age as a result of increasing sclerosis at the margin of the lesion, and never result in any of the potential complications (Fig. 1*A*). Cutaneous pigmentation is not usually associated with monostotic fibrous dysplasia.

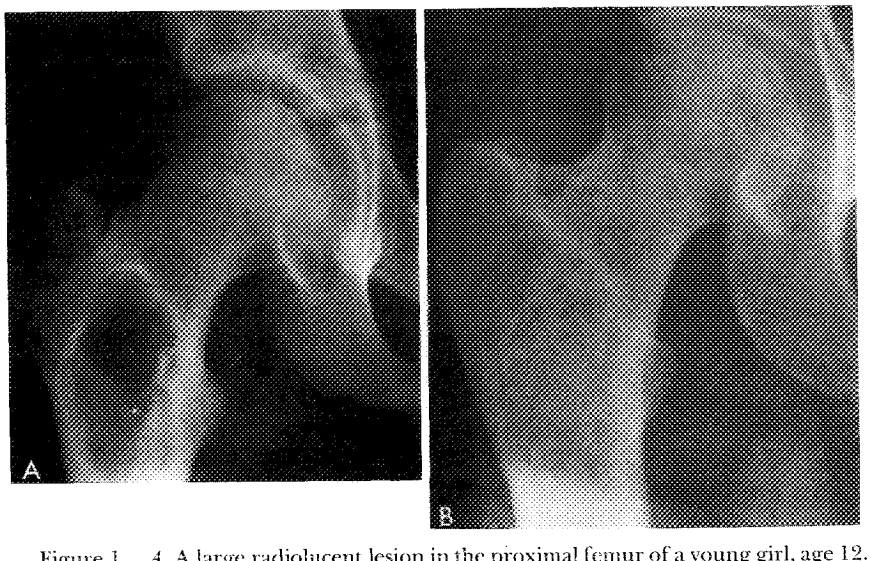

Figure 1. *A*, A large radiolucent lesion in the proximal femur of a young girl, age 12. Biopsy confirmed the lesion as showing fibrous dysplasia. Curettage and grafting were not performed. *B*, Radiograph ten years subsequently with marked healing of the lesion.

Polyostotic Lesions

At times all of an individual's skeleton may seem to be affected by fibrous dysplasia (Fig. 2). Twenty-five per cent of the patients with polyostotic fibrous dysplasia are reported to have more than 50 per cent of the skeleton involved at the time of initial presentation.[22] Two-thirds of these patients may have symptoms before the age of 10. On the other hand, there are cases in which a limited number of bones are involved. Segmental distribution of the lesions in a single extremity represents an important clinical and roentgenographic hallmark of the polyostotic form of fibrous dysplasia.[26] Extensive skeletal involvement does not seem to develop in patients who have mild disease at the onset. The more severe and earlier the skeletal involvement, the more rapidly the disease progresses with multiple fractures and deformities. In the review by Harris and his colleagues[22] from our hospital, over 85 per cent of the patients sustained fractures and 40 per cent had three or more. The lower extremity deformities and leg length discrepancies are secondary to femoral or tibial bowing, pathological fractures that heal with angular deformity, or epiphyseal growth changes. Although Albright and his associates[22, 32] originally stated that long bone involvement with fibrous dysplasia spared the epiphysis, epiphyseal involvement occurring prior to closure of the plate has been reported. What effect these epiphyseal lesions have upon growth or deformity is not clear. Craniofacial involvement is seen in half the patients with polyostotic fibrous dysplasia.

Fibrous dysplasia affecting the spine is quite rare. The lumbar portion of the spine is more commonly involved with vertebral collapse,

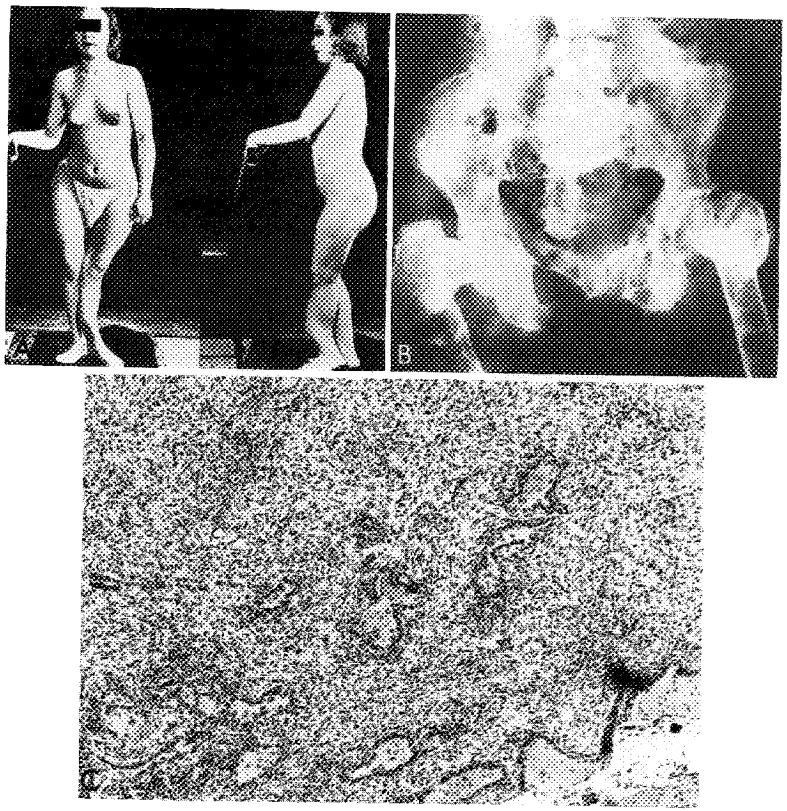

Figure 2. *A*, A 10 year old girl with Albright's syndrome. There is a large café au lait spot, which does not cross the midline anteriorly and follows the distribution of the skeletal lesions involving the left lower extremity. *B*, Radiograph of the pelvis. Fractures of the pubic rami on the left are recent. The right femur, which has not yet been treated, shows the typical shepherd's crook deformity. On the left side a similar deformity with an intracapsular fracture of the neck of the femur was treated by a high subtrochanteric osteotomy. *C*, Photomicrograph of tissue taken from the left proximal femur. The field is quite typical, except for the prominent osteoblasts rimming the trabeculae of woven bone (see text).

angular deformity, and rarely spinal cord compression. Eight instances of spinal cord compression have been documented.[32] Spinal cord compression is not always related to the vertebral collapse, but can be produced by a posteriorly expanding fibrous tissue mass.

Our impression is that puberty does seem to have a beneficial quieting effect on disease activity; this issue is not entirely resolved. Monostotic lesions, inactive after puberty, have become active again later in life.[23] Again, contrary to our personal experience (C.C.), Harris and associates[22] could find no effect of puberty in preventing occurrences of new lesions, reducing the incidence of fracture, or minimizing the progression of existing lesions. Pregnancy can be associated with reactivation of quiescent lesions.[23]

Albright's Syndrome

The rare syndrome characterized by osteitis fibrosa disseminata, areas of cutaneous pigmentation and endocrine dysfunction, and precocious puberty in females was thoroughly described by Albright and his associates[2] in 1937 (Fig. 2). Recent publications have expanded the original endocrine dysfunction to include cases of acromegaly, hyperparathyroidism, hyperthyroidism, and Cushing's syndrome.[5, 12, 14, 15, 21, 28] The pathogenesis of this intriguing syndrome remains unsettled. Hall and Warrick[20] proposed that the endocrine manifestations are due to hypersecretion of hypothalamic releasing hormones. A more recent report suggests that Albright's syndrome may represent an entity in which multiple organs initially or subsequently function in an autonomous fashion.[12] Although Albright's syndrome predominates in females, male involvement is not unknown. The syndrome may begin in infancy with vaginal bleeding in girls. Precocious menses (before nine years of age) is seen in about one-third to one-half of girls with the syndrome.[8] The major problems affecting these children are secondary to the various endocrinologic effects on body habitus with precocious sexual development, rapid bone maturation, early epiphyseal closure, and the ensuing short stature.[8] No adequate treatment exists for the premature epiphyseal closure that occurs.

All patients with polyostotic fibrous dysplasia and cutaneous pigmentation do not exhibit precocious puberty or other endocrine changes. Likewise the osseous lesions can occur with precocious puberty without cutaneous pigmentation.[14] However, the most common extraskeletal manifestation of fibrous dysplasia is abnormal cutaneous pigmentation. The pigmentary changes are generally described as multiple, clearly demarcated, flat melanotic areas that stop at the midline and have irregular dented margins. The skin texture of the pigmented area is exactly the same as that of the surrounding normal skin. The most frequent locations of these pigmented areas are the sacrum, buttocks, and upper spine.[2] The distribution has a unique tendency to parallel the distribution of the bone lesion in the skeleton. These pigmented macules have always been a cause of confusion in differentiating between neurofibromatosis and fibrous dysplasia. Albright offered a distinction in that the lesions in fibrous dysplasia with their irregular and dented margins are likened to the coast of Maine, in contrast to the irregular smooth outline of the pigmented areas in neurofibromatosis, which are likened to the coast of California.[8] Clinically the color and configuration have not been distinctive.

Conventional microscopy has failed to distinguish any difference between the macules found in either of the two conditions. There has been recognition of increased melanocyte activity with melanin deposition in the macules. A combined skin splitting and histochemical reaction has demonstrated giant pigment granules in malpighian cells or melanocytes or both in normal skin and in all tested patients with neurofibromatosis, but in only 10 per cent of patients with Albright's syndrome.[4] This would suggest that there may be a means of providing

a differentiation between the two conditions. In most instances it is not possible to make a diagnosis of either condition on the basis of a single macule. The presence of a diffuse, freckle-like pigmentation in the axilla is characteristic of neurofibromatosis, when present, and has not been seen in Albright's syndrome.[4] Nothing has been revealed to explain the relationship of the pigmentation to the various features of the syndrome. Observation of the pigmented macules at birth should not be casually dismissed, since these lesions have preceded the development of sexual precocity in fibrous dysplasia.

RADIOLOGIC FEATURES

The osseous lesions of monostotic fibrous dysplasia, isolated from other clinical features, are not radiologically distinctive (Figs. 1*A*, 3*A*). When several bones are affected, with or without adjacent axial lesions, the diagnosis becomes more apparent (Fig. 2*B*). The typical lesion is a large radiolucent area that has been frequently mislabeled as "cystic." The lesion in reality is not cystic but contains solid masses of fibro-osseous tissue. Whether the lesion is radiolucent or has a ground glass appearance depends upon the amount of osseous material within the lesion. The skeletal changes vary with the location of the lesion. In extremity lesions, cortical thinning, erosion, and scalloping from within, expansion, or enlargement of the bony contour occurs. These changes may involve a segment or the entire extent of a bone. The periosteal surface of the bone is not disturbed unless it is involved by a fracture. Fractures heal within normal time periods with a collar of callus. Increased density at the base of the skull, thickening of the occiput, obstruction of the paranasal and nasal sinuses, and displacement in the orbital cavity are characteristic changes involving the skull. A bone scan can be helpful to indicate the activity of an individual lesion, the extent of skeletal involvement, and sometimes recurrence postoperatively.[19]

Extreme hyperemia and marked uptake on the delayed image can present difficulties in differentiating the lesion of fibrous dysplasia from a malignant tumor, osteomyelitis, or bone trauma with a subperiosteal reaction. In those instances the diagnosis must be made in correlation with the plain radiographs. The differential diagnosis of the monostotic lesion must encompass: a solitary bone cyst, enchondroma, nonossifying and ossifying fibromas, enchondromatosis, and hyperparathyroidism (Fig. 1*A*). The histologic examination can only discern enchondroma from fibrous dysplasia, although both may be seen together. The clinical and chemical examinations can distinguish hyperparathyroidism. The alkaline phosphatase level is seldom elevated (except with a fracture) in fibrous dysplasia and never the serum calcium level. In enchondromatosis the lesions may be confined to one side of the body, as seen with fibrous dysplasia. The clinical and radiographic appearance of neurofibromatosis may resemble that in fibrous dysplasia, but the histologic picture is quite distinctive.

PATHOLOGY

In the early experience with fibrous dysplasia, there were few comprehensive pathologic descriptions because of the scarcity of tissue available for study. Most surgical specimens are small fragments removed at biopsy, the involved bones are seldom amputated, and autopsy material is limited. On gross appearance, fibrous dysplasia involves a replacement of normal bone with masses of fibrous tissue, some of which may be gritty or sandy to touch. These masses of tissue are solid, except for small degenerative cystic areas containing fluid. The cystic areas are associated with cellular necrosis or areas of prior surgery. With fracture, periosteal callus and hemosiderin staining of the tissues appear.

Histologic study of a lesion shows a mature collagenous tissue in which are contained trabeculae of woven bone. Within a single lesion the histologic features can vary from mature connective tissue with sparse bony trabeculae to areas composed largely of osteoid material and woven bone. The younger the lesion and the less spindled the cells, the larger the nuclei and the less collagen is seen. The more mature lesions demonstrate a dense collagenization, and the cellular pattern is that of organized fibrous tissue seen elsewhere in the body. The fibrous tissue seen in fibrous dysplasia may be similar to the fibrous tissue seen in other lesions, such as desmoplastic fibroma and nonosteogenic fibroma. However, it is the appearance of the bone within the lesion that permits the diagnosis. The bony trabeculae are thin and irregular in shape and size, lack a specific tissue orientation, and are formed through osseous metaplasia of the surrounding fibrous tissue.

Under polarized light and with reticulum stains the bone is demonstrated to be woven in type rather than lamellar.[40] In the classic lesion there is a conspicuous absence of the osteoblasts rimming the trabeculae of woven bone that are so characteristic of osteoblastoma and ossifying fibroma. In the more aggressive lesions of fibrous dysplasia that are seen in younger children, osteoblasts rimming the woven bone spicules are commonly seen in our experience (Fig. 2*C*). This leads to a difficult differentiation of fibrous dysplasia from ossifying fibroma. However, ossifying fibromas are classified as osseous variants of fibrous dysplasia. The routine histologic features may be similar. Lamellar transformation of woven bone is seen in cases of ossifying fibroma. The lamellar appearance of appositional bone is not usual in fibrous dysplasia. Aside from the histologic study, the clinical and roentgenographic appearance of the lesion becomes important. The separation of these two entities has therapeutic consequences, since ossifying fibroma is an aggressive lesion that can be arrested only by wide excision, periosteal stripping, and bone grafting.[40]

The lesions of fibrous dysplasia are not avascular and blood vessels can be seen. Giant cells are sparse, except in areas of degeneration and hemosiderin deposition. Cartilage is present, although some believe it to be rare and present in only about 10 per cent of the lesions.[40] When carti-

lage is present in considerable amount, the radiographic and pathologic appearance can simulate that of a typical enchondroma.

This finding of cartilage within an area of fibrous dysplasia has consistently raised speculation concerning the inter-relationship between enchondromatosis and fibrous dysplasia. Cohen[9] has reported atypical patterns of fibrous tissue and foam cells in fibrous dysplasia. A relationship between fibrous dysplasia and adamantinoma is continually questioned. In all our personal (C.C.) cases of adamantinoma, fibrous dysplastic tissue was prominent. However, Huvos and Marcove could not demonstrate the microscopic presence of any intracortical fibrous dysplasia in a series of 14 cases of adamantinoma.

Repeated biopsy examinations have been performed at random intervals in several patients to study the histologic evolution of the bone lesions.[22] With the passage of time there were only very subtle changes—a decrease in cellularity, an increased deposit of collagen, and a slight decrease in the amount of bone relative to fibrous tissue. None of the woven bone trabeculae underwent conversion into a lamellar bone pattern. Resorption and accretion of bone occurred, but the remodeling did not result in an orientation of the trabeculae along stress lines. One interesting feature was the apparent discrepancy between the large amount of new bone formation seen in serial biopsy specimens from the same region in a given patient over a span of five to 10 years and the finding that none of the lesions became more sclerotic, either histologically or radiographically, as time passed.

MALIGNANT TRANSFORMATION

Histologic examination in fibrous dysplasia demonstrates a uniformly benign appearance of the fibrous tissue with no evidence of mitoses or cellular atypism. As of 1942 there were no reported instances of malignant degeneration occurring in areas of fibrous dysplasia,[26] and subsequent statistical information concerning the frequency of malignant transformation has been imprecise and misleading. The actual incidence is most likely less than 1 per cent. At the present time there are less than 50 reported cases of malignant transformation occurring in fibrous dysplasia.[30] Some of those cases are in question owing to the lack of adequate documentation of the pathologic diagnoses. Less than half the cases occurred in patients with polyostotic fibrous dysplasia. In patients presenting initially with an obvious malignant skeletal tumor, it is easy to overlook the small foci of a background lesion, unless the pathologist specifically searches for them.

The differentiation of the malignant transformation can vary. Fibrous dysplasia is a mixture of bone forming connective tissue with fibrous and chondroid elements. Therefore, a variety of directions, depending on the types of tissue in the lesion, are available for malignant transformation. The most frequent type described is osteosarcoma. Fibrosarcoma, chondrosarcoma, and giant cell tumors have been reported. The facial bones and femur are the most commonly affected

sites undergoing malignant transformation.[10] Although prior surgical intervention did not seem to be a factor in precipitating malignant change, malignant transformation has occurred following irradiation of previously benign lesions of fibrous dysplasia.[16] There does not seem to be a specific sex predilection or age group distribution. A reported five year survival rate of 52 per cent indicates that an aggressive approach should be directed to eradication of the lesion.[24] There is no apparent way to explain why this malignant transformation occurs. Everyone agrees that malignant transformation in fibrous dysplasia is more common than previously appreciated.

ORTHOPEDIC CONSIDERATIONS

No medical therapy can effectively halt the slowly progressive lesions of polyostotic fibrous dysplasia. Various hormonal agents are under continued investigation in Albright's syndrome. Radiation therapy has no proven efficacy and may induce malignant transformation. The orthopedist must assume the practical problems of diagnosis, fracture management, reconstruction for deformity, and attempts at tumor therapy. The age of the patient, location and size of the lesion, and the biologic behavior of fibrous dysplasia influence our management decisions.

"The mere fact of their [skeletal lesions] presence is not an indication for treatment"[25] reinforces our personal viewpoint that the lesions are surgically overtreated. Monostotic lesions are operated upon to determine a diagnosis and eradicate the lesion, whereas polyostotic fibrous dysplasia requires treatment of fracture and deformity. Biopsy should not always necessitate curettage and grafting, if one has prior experience in treating such lesions. The lesion depicted in Figure 1*A* was subjected to biopsy examination but not curetted or grafted. Over a period of 10 years the lesion has healed (Fig. 1*B*). The area of biopsy should be well placed and not too large. The hazards of biopsy and curettage of isolated extremity lesions are pathologic fracture and recurrence. There is no available information that accurately indicates the success rate with curettage and grafting of the lesions of fibrous dysplasia.

The choice of graft materials varies with the individual surgeon. Some prefer autograft; others, freeze dried corticocancellous homografts; a few recommend allograft. Those arguing against the autograft, except in small single lesions in older children, point out correctly that the autograft material is limited, the lesions generally require large amounts of graft material, the pelvis may be involved with the disease itself, and the frequency of recurrence is probably not influenced by the type of graft material used. Cortical and allograft materials add structural support and require a longer period for resorption. With these graft materials there is a delay in the true appearance of a recurrence. We believe that any and all types of material can be

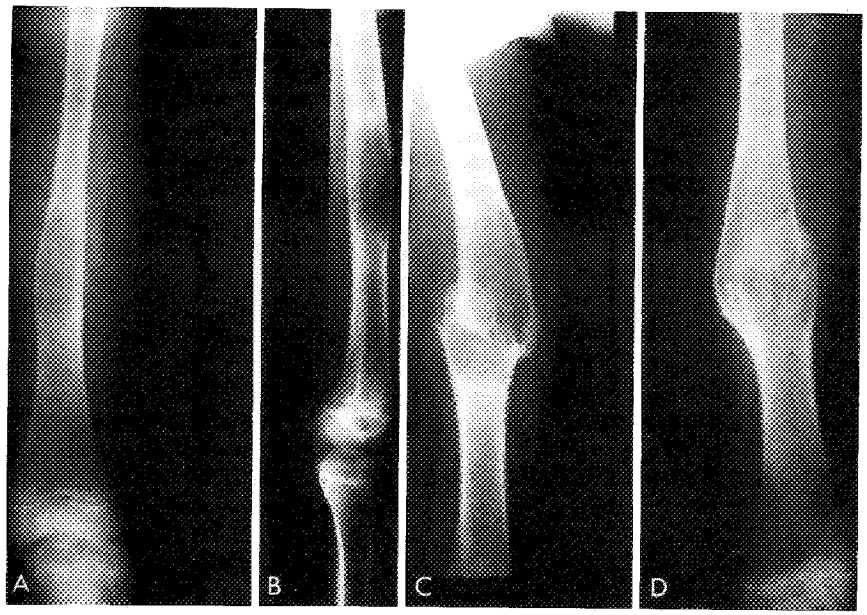

Figure 3. *A* and *B*, Pathological fracture of the left femoral diaphysis through an area of fibrous dysplasia. *C* and *D*, Twelve weeks later. The patient was treated in skeletal traction for three weeks and then allowed ambulation in a functional orthosis.

justified on the basis of filling any empty osseous cavity. In spite of possible recurrence, successful obliteration of the lesion can be achieved. Excisional surgery is preferred for superficial lesions, e.g., those of the ribs and clavicles. Finger and hand lesions are appropriate for subtotal excision and grafting.

The major skeletal effect of fibrous dysplasia is to weaken the structural integrity of individual bone units. With any brittle or soft bone disorder, the bone deforms with stress; the results are fracture and deformity. Fractures in fibrous dysplasia are produced frequently by mild trauma; they seldom become displaced and appear to heal at a normal rate. The management of fractures is standard according to the site of the fracture.

Traction techniques are best for diaphyseal fractures of the femur (Fig. 3). Upper and lower extremity cast bracing has reduced the period of traction and nonfunctional immobilization in these children. We are now able to shorten the overall period of hospitalization and allow them to be active in home and school activities with functional bracing.

The problem area of treatment remains the proximal femur. After closure of the proximal epiphyseal plate, femoral neck lesions can be treated with curettage, grafting, and nail-plate fixation. Whenever a fracture has occurred, reduction of the proximal fragment into the valgus position is critical to secure stable fixation, compensate for any leg length discrepancy, and prevent recurrent deformity. Fractures of the femoral neck with a varus deformity can be secured in situ with a

lag screw; then an intertrochanteric valgus osteotomy is performed below to achieve a stable fixation in valgus, which is held with an appropriate nail-plate device. Too often there is a tendency to accept a less than optimal position in patients with extensive skeletal disease. Advanced femoral head and neck deformities in polyostotic fibrous dysplasia may require proximal resection for relief of pain and restoration of motion. There are no known instances of prosthesis replacement in such circumstances. The principles of fracture management in fibrous dysplasia are not unique but are common to all patients, namely, restoration of alignment and function with an emphasis on functional bracing techniques when applicable.

The earlier and more severe the extent of bony involvement, the more deformities will appear. Upper extremity deformities are less frequent. The weight bearing portions of the skeleton, spine, and lower extremities bear the burden of deformity. Back pain with vertebral collapse should be actively treated. A light weight orthosis should be utilized to prevent deformity after appropriate periods of bed rest for comfort. Long term spinal bracing may be necessary in the growing child to prevent severe kyphosis. The spectrum from mild coxa vara to the severe shepherd's crook deformity is seen in advanced and untreated cases involving the proximal femur. Early osteotomy with supplemental nail-plate fixation is necessary. In some circumstances early femoral and tibial diaphyseal osteotomies and intramedullary fixation are necessary to control the severe femoral and tibial bowing. Serial osteotomies may be necessary if the deformities occur early and are progressive. A variety of internal fixation devices can be used as adjuncts to the osteotomy. There are problems with any fixation device in a soft bone disorder. Refracture below the plates is common if the plate does not extend beyond the diseased area of bone. Screw fixation into thin cortices is poor, and the use of a nut and bolt combination can help in securing the plate to the diaphysis.

Leg length discrepancies are secondary to structural deformities and growth disparities of the extremity. Correction of the deformity or epiphyseal arrest on the opposite side must be individualized for each patient and requires good judgment on the part of the orthopedist.

As mentioned, the lesions of fibrous dysplasia are not painful in themselves. Complications of the lesion produce pain. A painful lesion signals impending fracture, enlargement, or a malignant change. The authors have had no personal experience with malignant transformation in fibrous dysplasia. The experience of others indicates that serious consideration should be given to aggressive radical local resection or amputation as indicated by specific circumstances.

SUMMARY

An overview of the spectrum and natural history of fibrous dysplasia is an essential prerequisite of management. Current medical interest has focused upon the intriguing pathogenesis of the bone lesions, the

endocrinopathies, the cutaneous pigmentation, and malignant transformation. The mundane aspects of diagnosis and the complications of a soft bone disorder reside with the clinical orthopedist. Diagnosis is straightforward with the aid of biopsy. Conservatism in the surgical treatment of monostotic lesions and experience in the care of fractures and deformity in polyostotic fibrous dysplasia constitute the lessons learned in a series of over 50 patients with fibrous dysplasia.

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